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Extra-hepatic portal vein aneurysm: A case report, overview of the literature and suggested management algorithm

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ABSTRACT

INTRODUCTION: Portal venous aneurysms are a rare finding. The reported incidence is on the rise with increasing use of modern imaging techniques in clinical practice. However, there is still much to be elicited regarding their aetiology, natural history, and management.
PRESENTATION OF CASE: An 80-year-old woman presented with abdominal pain and nausea. Investigations showed a hypoechoic area in the region of the head of pancreas on ultrasound, which was found to be a portal venous aneurysm on CT. In view of her multiple comorbidities, a conservative approach was taken.
DISCUSSION: Portal venous aneurysms represent approximately 3% of all venous aneurysms with a reported prevalence of 0.43%. They may be congenital, due to failure of complete regression of the right vitelline vein, or acquired secondary to portal hypertension. The primary presentation of portal vein aneurysm is abdominal pain, followed by incidental detection on imaging, with a minority of patients presenting with gastrointestinal bleeding. Complications of PVA include thrombosis, biliary tract obstruction, inferior vena cava obstruction, and duodenal compression. On the whole PVAs are stable and have a low risk of complications with 88% of patients showing no progression of aneurysm size or complications on subsequent follow up scans.
CONCLUSION: We recommend that portal venous aneurysms be assessed using colour Doppler ultrasonography in the first instance with CT scans reserved for indeterminate cases or symptomatic patients. Due to the slow progression of such aneurysms, surgery is recommended only for symptomatic patients or those with complications secondary to portal venous aneurysms.

1. Introduction

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and splenic vein (SV) in the region of spleno-portal junction. They are rare with less than 200 cases reported in the literature since it was first described in 1956.[1] Although it is being identified more frequently, the aetiology, natural history and management choices are still relatively unclear. Herein we present a case of an incidental portal venous aneurysm discovered on routine investigations for suspected gallstone disease. Subsequently, a review of the literature is presented with a particular focus on the management options for this increasingly recognised condition.

2. Case report

appetite remained good. Her past medical history included angina requiring a double coronary artery bypass graft, intermittent claudication, hypercholesterolemia and hypothyroidism. She had no significant history of previous abdominal pathology.

An abdominal ultrasound (US) scan showed a thin-walled gallbladder with multiple gallstones and a normal common bile duct. A hypoechoic area was noted in the region of the head of the pancreas initially thought to represent a cystic mass. A subsequent contrast enhanced computed tomography scan (CT) showed aneurysmal dilatation at the confluence of portal vein, SMV and SV measuring 42 mm × 40 mm × 37 mm (Figs. 1 and 2). There were no features of thrombosis, portal hypertension, chronic liver disease, pancreatic mass or pancreatitis. No aneurysmal change was noted in the arterial tree. Laboratory tests including the liver function tests were normal apart from showing mild renal impairment.

In view of her multiple co-morbidities and symptoms attributable to gallstone disease, a conservative approach was

Abbreviations: PVA, portal vein aneurysm; PV, portal vein; SV, splenic vein; SMV, superior mesenteric vein.

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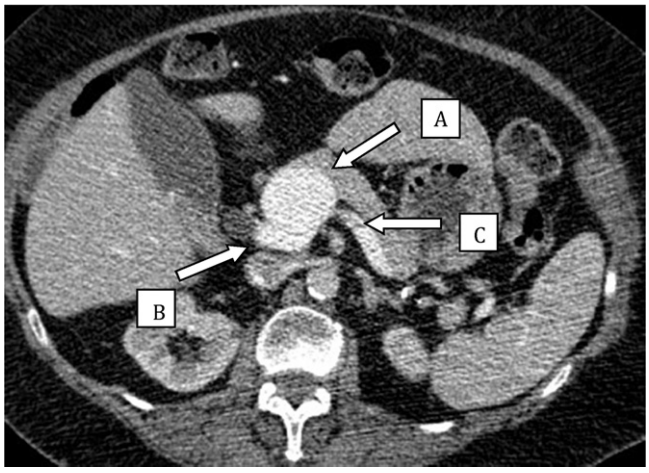

Fig. 1. Axial CT shows portal vein aneurysm (A) and the confluence of main PV (B), splenic vein (C).

adopted with regards to the incidentally detected portal venous aneurysm.

3. Discussion

Primary venous aneurysms are much less common compared to arterial aneurysms. However, reported incidence of PVA has increased in recent years due, most likely, to the increased availability of advanced imaging techniques in clinical practice. They represent approximately 3% of all venous aneurysms[2] with a reported prevalence of 0.43%.[3] First reported by Barzilai and Kleckner,[1] there are now over 170 cases described in the literature.[4–7] The majority of these are located in the main extra-hepatic PV with aneurysms of the SV-SMV confluence. PVAs may be either congenital or acquired but the exact aetiology still remains controversial and often difficult to determine. Portal hypertension secondary to chronic liver disease is considered the most common cause of acquired portal vein aneurysms.[2,8] Portal hypertension leads to intimal thickening with compensatory medial hypertrophy. Progressive replacement of the medial hypertrophy by fibrous tissue weakens the tensile strength of the venous wall making it more susceptible to aneurysmal dilatation.[8] Other causes described in the literature include pancreatitis, trauma and previous surgical intervention.[7,9,10] However, for many cases, there is no obvious underlying disease process attributable as the cause of PVA. Embryologically, the portal venous system develops from the vitelline and umbilical veins, which are responsible for venous drainage of the intestine.[11] Supporters of a congenital aetiology of portal vein aneurysms suggest that failure of a complete regression of the right vitelline vein may leave a diverticular remnant that can subsequently enlarge to form a saccular aneurysm later in life.[9,12,13] A report of an in-utero diagnosis of portal vein diverticulum lends support to this theory.[14] Other hypotheses attribute formation of portal vein aneurysms to an inherent weakness in the venous wall.[15]

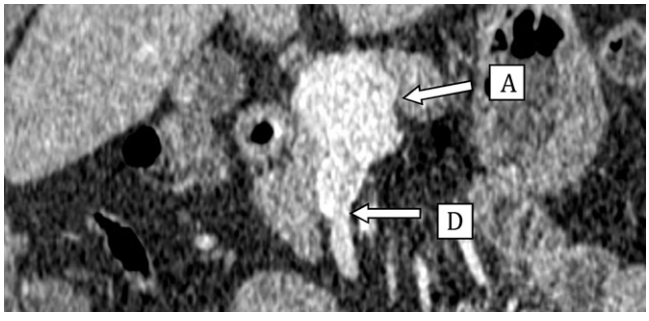

Fig. 2. Coronal reformatted CT image shows portal vein aneurysm (A) at the confluence with SMV (D).

Several US and autopsy studies have shown that PV size varies considerably in normal population.[16–18] A study by Doust and Pearce reported a maximum diameter of 1.5 cm in normal patients rising to 1.9 cm in cirrhotic patients.[18] Therefore, a focal dilatation of greater than 2 cm is now considered diagnostic in the evaluation of extra-hepatic PV aneurysms.[14,18] An anechoic cyst-like lesion near the portahepatis is the characteristic finding on B-mode ultrasonography[9] with Doppler or colour flow studies helping to confirm the diagnosis by showing blood flow through the lesion, thus differentiating it from a liver cyst. Contrast enhanced CT and Magnetic Resonance Angiography (MRA) are useful adjuncts in patients with equivocal US findings or for anatomical characterisation of the aneurysm when planning for surgical interventions.

The most common presentation of portal vein aneurysms is abdominal pain (44.7%), followed by incidental detection on CT and US (38.2%), with a minority of patients presenting with gastrointestinal bleeding (7.3%).[4] Complications of PVA include thrombosis,[1,3,19–21] biliary tract obstruction,[15] inferior vena cava obstruction,[22] and duodenal compression.[22,23] On the whole PVAs are stable and have a low risk of complications with 88% of patients showing no progression of aneurysm size or complications on subsequent follow up scans.[4,22,24] Indeed, there is a recent report of a 6 cm saccular aneurysm undergoing spontaneous complete regression over many years of follow up.[6] Rupture of a PVA has been reported in three cases, of which one patient died.[1,20,25] Symptomatic or expanding aneurysms are generally considered indications for surgical intervention. The type of surgical intervention depends on associated features. Patients with portal hypertension and portal vein thrombosis have usually undergone shunt surgery.[24] This surgery aims primarily to decompress the portal venous system rather than treat the aneurysm itself although reduced pressures could possibly prevent progressive dilatation of the aneurysm. For patients without portal hypertension, the operation of choice is aneurysmorrhaphy. By resecting out redundant parts of the venous wall and re-suturing, portal circulation is preserved and laminar blood flow restored thus reducing the risk of future thrombosis.[22]

The natural history of PVAs is not fully understood and there is no consensus for their management. We propose the following algorithm to help guide management decisions. PVAs should initially be assessed by colour Doppler ultrasonography with CT scans reserved for indeterminate ultrasound scans and symptomatic patients. CT scans would serve a dual purpose to both rule-out any other concurrent pathology and to help delineate anatomy. Patients that are asymptomatic could be managed conservatively with serial US imaging. Symptomatic patients with severe abdominal pain, symptoms of pressure effect, or with expanding aneurysms and/or complications such as thrombosis or rupture would require surgical intervention. If the aneurysm is associated with portal hypertension, portal-venous shunts should be considered in the first instance. For all other aneurysms without concomitant portal hypertension or cirrhosis, primary aneurysmorrhaphy should be attempted (Fig. 3). Patients with thrombosis that extends from the main portal vein into the splenic or superior mesenteric veins should also undergo thrombectomy if possible. The management of large aneurysms in asymptomatic patients is still controversial and should be decided on a case-by-case basis with discussion with the patient about the risks and benefits. Further studies are required to assess the long-term risk of complications in this population group.

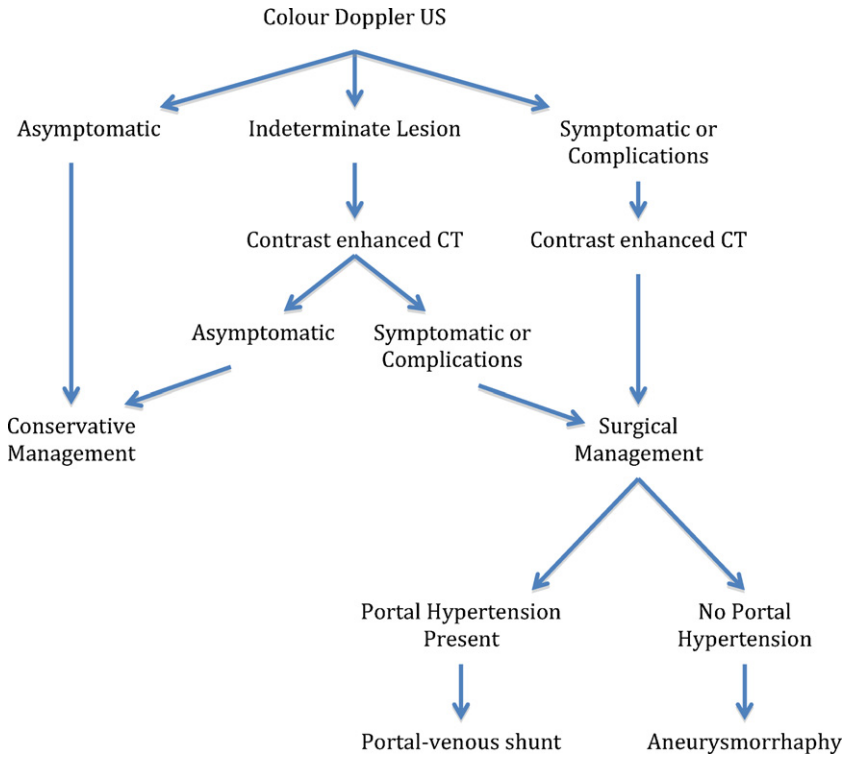

Fig. 3. Proposed management algorithm. Patients should be initially assessed using Doppler ultrasonography with CT scans reserved for indeterminate lesions and patients with symptomatic PVA. Surgical management should be considered in symptomatic patients with choice of surgery dependent of the presence of portal hypertension. Conservative management should be taken for asymptomatic patients. Patients with large aneurysms should be considered on a case-by-case basis.

In summary, extra-hepatic portal vein aneurysms are a rare occurrence but one that is becoming more widely recognised with the increased use of CT and MRI in clinical practice. The portal vein aneurysm in our patient is likely to be congenital in origin, as she had no signs of liver disease or portal hypertension. Our management choice of expectant treatment is in keeping with the evidence in the literature.

Conflicts of interest

The authors report no conflicts of interest.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Asif Jah identified the case report. The literature search was performed by Ruichong Ma. Specialist radiological input was provided by Teik Choon See. The case report was written by Ruichong Ma and reviewed and edited by Anita Balakrishnan, Siong Seng Liau, Raaj Praseedom and Asif Jah. The guarantor of the work is Mr Asif Jah.

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